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Twenty-year analysis of the rarely diagnosed nutcracker syndrome

Nadir görülen nutcracker sendromunun yirmi yıllık analizi

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ABSTRACT

Aim: Nutcracker syndrome is a very rare cause in patients presenting to the emergency department with abdominal pain. Early consideration in differential diagnosis will reduce the time spent for diagnosis and morbidity, as well as provide the correct treatment. We aimed to contribute to the literature by determining the clinical relations of these cases with laboratory, imaging and treatment data.

Material and Method: Twenty-seven patients over the age of 16 who presented to the emergency department with abdominal pain between January 2000 and December 2020 were included in this study. The ages of the patients were between 16-39 and the mean was 25.19±7.00 years. Demographic characteristics, clinical findings, laboratory parameters, radiological evaluations, and treatment modalities of patients were evaluated retrospectively.

Results: Abdominal pain and flank pain were the most common reasons for admission to the emergency department of 27 patients included in the study. In the anterior group, 17 (77%) patients had flank pain, 10 (45%) patients had dysmenorrhea, 11 (50%) hematuria, 10 (45%) proteinuria, and 9 (40%) patients hematuria and proteinuria. In the posterior group, there was no hematuria, proteinuria, anorexia, nausea and vomiting, and oral intolerance. In cases with anteior nutcracker syndrome, 12 (54%) doppler ultrasonography, 13 (59%) computed tomography, 5 (22%) magnetic resonance imaging were performed. Conservative treatment was applied to 11 (40%) patients in the anterior group and 3 (11%) patients in the posterior group. Endovascular surgery was performed on 5 (22%) female patients.

Conclusion: Nutcracker syndrome should be investigated in adult patients who present to the emergency department with abdominal pain and whose diagnosis is unclear. Early diagnosis is critical in terms of treatment and morbidity.

Keywords: Emergency department, abdominal pain, nutcracker syndrome, hematuria, proteinuria

ÖZ

Amaç: Acil servise karın ağrısı ile başvuran hastalarda nutcracker sendromu çok nadir ortaya çıkan bir nedendir. Ayırıcı tanıda erken dönemde akla getirilmesi, hastanın tanı için geçirdiği süreyi ve morbiditeyi azaltacağı gibi, doğru tedaviyi uygulamayı da sağlayacaktır. Literatür eşliğinde bu olguların değerlendirilmesi amaçlandı.

Gereç ve Yöntem: Bu çalışmaya, Ocak 2000-Aralık 2020 tarihleri arasında acil servise karın ağrısı nedeniyle başvuran 16 yaşından büyük 27 hasta dahil edildi. Hastaların yaşları 16 ile 39 arasındaydı ve ortalaması 25.19±7.00 yıldı. Bu hastaların demografik özellikleri, klinik bulguları, laboratuar parametreleri, radyolojik değerlendirmeleri ve tedavi şekilleri retrospektif olarak değerlendirildi.

Bulgular: Çalışmaya dahil edilen 27 olgunun acil servise en sık başvuru nedeni karın ağrısı ve yan ağrısıydı. Anterior grupta 17 (%77) hastada yan ağrısı, 10 (%45) hastada dismenore mevcutken, 11 (%50) hematüri, 10 (%45) proteinüri ve 9 (%40) hematüri ve proteinüri olgusu vardı. Posterior grupta hematüri, proteinüri, iştahsızlık, bulantı ve kusma ve oral intolerans yoktu. Anterior nutcracker sendromu grubunda 12 (%54) Doppler ultrasonografi, 13 (%59) bilgisayarlı tomografi, 5 (%22) manyetik rezonans görüntüleme yapıldı. Anterior grupta 11 (%40) hastaya, posterior grupta 3 (%11) hastaya konservatif tedavi uygulandı. 5 (%22) kadın hastaya endovasküler cerrahi uygulandı.

Sonuç: Erişkin dönemde acil servise karın ağrısıyla başvurun ve tanısı geciken olgularda nutcracker sendromu düşünülmelidir. Erken tanı değerlendirmesi; tedavi ve morbidite açısından önem taşımaktadır.

Anahtar Kelimeler: Acil servis, karın ağrısı, nutcracker sendromu, hematüri, proteinüri

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INTRODUCTION

In 1950, El Sadr and Mina demonstrated that the left renal vein was compressed between the abdominal aorta and superior mesenteric artery (1). De Schepper provided Nutcracker syndrome (NCS) its name in 1972 (2). It is characterized by compression of the left renal vein, however there are anatomical variants. At this level, it is defined as the narrowing of the left renal vein due to external compression and dilatation due to pressure increase before this segment. It is rarely diagnosed because there are no clear diagnostic criteria, the symptoms can show up in different ways, and it is not in the list of possible diagnoses. The Nutcracker Phenomenon defines the morphological findings due to compression of the left renal vein. It differs from NCS by the absence of clinical findings of the patient (3).

In the classification, NCS is defined in two main groups as anterior and posterior. The more prevalent anterior variant is caused by compression of the left renal vein between the aorta and the superior mesenteric artery (SMA). The posterior variant is rarer and results from compression of the retro aortic left renal vein between the aorta and vertebrae (4).

The most important point in diagnosis is to suspect this syndrome. In the laboratory, hematuria and proteinuria are directive findings. The imaging method to be preferred first is doppler ultrasonography (DUS), and retrograde venography is the gold standard in diagnosis. However, its invasiveness restricts its practical application. Other examinations include computed tomography (CT) angiography and magnetic resonance imaging (MRI) angiography (5). Treatment options are determined by the severity of the symptoms and include conservative and various surgical approaches. Conservative option comes to the fore, especially in children. Surgical methods include open surgery, laparoscopic surgery, and endovascular interventions (6).

In this study, we evaluated at 27 patients who were diagnosed with nutcracker syndrome and analyzed their clinical symptoms, laboratory results, imaging, and treatment options. We aimed to emphasize the clinical importance of the disease and to contribute to the clinical, diagnostic, and treatment management process.

MATERIAL AND METHOD

The study was carried out with the permission of Bağcılar Training and Research Hospital Clinical Researches Ethics Committee (Date: 15.01.2021, Decision No: 2021.01.1.01.001). All procedures complied with the ethical norms of the institutional and national committees responsible for human experiments and the Helsinki Declaration.

Due to the retrospective design of the investigation, no written informed permission forms were acquired from patients. The corresponding author certifies, on behalf of all authors, that there are no conflicts of interest.

Study Design and Population

Patients diagnosed with NCS admitted to the emergency department between January 2000 and December 2020 were retrospectively analyzed using the electronic medical archive system. Patients older than 16 years old who presented to the emergency department due to abdominal pain and were diagnosed with NCS were included in the study. Patients' age, gender, clinical symptoms and physical examination findings, laboratory and radiological imaging findings, affected sides, surgical findings, methods, procedures, and examination results were recorded.

The cases were grouped as anterior nutcracker syndrome (ANCS) and posterior nutcracker syndrome (PNCS). The patients' white blood cell (WBC), mean corpuscular volume (MCV), mean corpuscular hemoglobin (MCH), mean corpuscular hemoglobin concentration (MCHC), red cell distribution width (RDW), mean platelet volume (MPV), neutrophil (NEU), lymphocyte (LYM), neutrophil/lymphocyte ratio (N/L), glucose (GLC), alkaline phosphatase (ALP), alanine aminotransferase (ALT), aspartate aminotransferase (AST), lactate dehydrogenase (LDH), creatine kinase (CK), gamma glutamyl transferase (GGT) C-Reactive Protein (CRP) values were determined. Clinical symptoms and findings were grouped as side pain, abdominal pain, hematuria, proteinuria, hematuria+proteinuria (HP), side pain+hematuria (SPH), side pain + hematuria + proteinuria (SPHP), varicocele, pelvic congestion, dyspareunia, dysmenorrhea, intolerance, dyspepsia, anorexia, nausea, vomitting, weight loss. The cases were classified as renal doppler ultrasonography (RDUS), computed tomography, magnetic resonance imaging, retrograde venography (RV) according to imaging methods. Treatment conditions were grouped as conservative, surgical, and endovascular.

Statistical Analysis

The data obtained from the study were collected from the hospital's electronic archive system. This study's data were analyzed using the SPSS 20 (SPSS Inc., Chicago, IL, USA) software platform. Descriptive statistics were presented as mean±standard deviation or median (minimum-maximum) for continuous variables and as the number of cases and percentage (%) for nominal variables. Other data were analyzed using Microsoft Excel and simple descriptive statistics.

RESULTS

The study included 27 patients diagnosed with NCS. Sixteen of the patients were female and 11 were male. The ages of the patients were between 16 and 39 and the mean age was 25.19±7.00 years. Of the 22 ANCS patients, 14 were female, the mean age was 25.50±7.44 years, while two of the five PNCS patients were female and the mean age was 23.80±5.02 years. ANCS was common in women, while PNCS was common in men. Mean hemogram parameter values of all cases, WBC 9.49±2.95 (10^3/uL), MCV 85.11±10.77 (fl), RDW 14.30±1.22 (%), NEU 7.85±2.48 (10^3/uL), LYM 2.05±0.84 (10^3/uL) µL), and the NLR was 4.95±3.86. The mean biochemistry values are GLC 103.67±13.28 mg/dL, amylase 85.33±19.41 (U/L), ALP 55.89±15.24 (U/L), ALT 23.00±13.99 (U/L), LDH 198.74±35.10 (U/L)), CK 146.41±75.32 (U/L), GGT 34.52±12.01 (U/L), CRP 4.62±2.78 mg/L. The average of all biochemistry values evaluated was within normal limits. In the ANCS group, the neutrophil count was 7.57±2.56 (10^3/UL), NLR was 4.32±2.96, and the creatine kinase was 141.09±71.99 (U/L). In the PNCS group, these values vere 9.07±1.81 (10^3/UL), 7.71±6.27, and 169.80±93.91 (U/L) respectively. In addition, ALP values differed as 57.45±15.55 U/L in ANCS and 49.00±12.92 U/L in PNCS. (**Table 1**).

Side pain was present in 17 (63%) of 22 (81.5%) cases, and in 2 (7.4%) of 5 (18.5%) PNCS cases. Abdominal pain was observed in 15 (55.6%) cases in the ANCS group and in 4 (14.8%) cases in the PNCS. There were 11 (40.7%) hematuria, 10 (37%) proteinuria, 9 (33.3%) HP, 8 (29.6%) SPH, 7 (25.9%) SPHP cases in the ANCS group. Hematuria and proteinuria were not found in any of the PNCS cases. Common complaints were dyspareunia 7 (25.9%), dysmenorrhea 10 (37%), and dyspepsia 11 (40.7%) in the ANCS group. Varicocele, pelvic congestion, dyspareunia, dysmenorrhea, and dyspepsia were observed in 1 (3.7%) case in the PNCS group. Side pain was common in women and abdominal pain in men. Hematuria and proteinuria were observed more frequently in female cases. On the other hand, weight loss was also higher in female cases (**Table 2**).

Table 1. The relationship of nutcracker syndrome subtype and gender groups with baseline characteristics and laboratory values

NCS	All Patients n: 27	NCS Classification		Gender	
		ANCS n:22	PNCS n:5	Female n:16	Male n:11
	Mean±SD	Mean±SD	Mean±SD	Mean±SD	Mean±SD
Baseline Characteristics					
Age (year)	25.19±7.00	25.50±7.44	23.80±5.02	24.81±7.66	25.73±6.23
Female (n) / Male (n)	16/11	14/8	2/3	-	-
Hemogram Parameters					
WBC (10^3/uL)	9.49±2.95	9.46±3.00	9.62±2.91	10.33±3.15	8.26±2.22
MCV (fL)	85.11±10.77	85.31±11.72	84.20±5.67	87.43±4.15	81.73±15.96
MCH (pg)	30.07±2.44	30.25±1.36	29.26±5.68	30.05±1.50	30.10±1.43
MCHC (g/dL)	33.66±1.29	33.65±1.29	33.72±1.41	33.58±1.35	33.79±1.24
RDW (%)	14.30±1.22	14.12±1.04	15.10±1.72	14.41±1.39	14.14±0.96
MPV (fl)	8.46±0.95	8.31±0.94	9.08±0.78	8.38±1.05	8.57±0.81
NEU (10^3/uL)	7.85±2.48	7.57±2.56	9.07±1.81	8.31±2.71	7.18±2.03
LYM (10^3/uL)	2.05±0.84	2.09±0.79	1.84±1.14	1.83±0.81	2.36±0.83
N/L	4.95±3.86	4.32±2.96	7.71±6.27	5.48±3.36	4.18±4.55
Biochemistry Parameters					
GLC (mg/dL)	103.67±13.28	103.54±13.88	104.20±11.61	104.88±13.42	101.91±13.52
Amylase (U/L)	85.33±19.41	85.82±18.09	83.20±26.94	85.56±24.06	85.00±10.57
ALP (U/L)	55.89±15.24	57.45±15.55	49.00±12.92	57.25±14.36	53.91±16.96
ALT (U/L)	23.00±13.99	23.68±14.96	20.00±9.11	22.75±16.45	23.36±10.11
AST (U/L)	24.15±9.25	24.37±9.83	23.20±6.91	22.94±10.74	25.93±6.62
LDH (U/L)	198.74±35.10	199.23±38.38	196.60±16.38	199.31±43.94	197.91±17.47
CK (U/L)	146.41±75.32	141.09±71.99	169.80±93.91	147.50±67.54	144.82±88.89
GGT (U/L)	34.52±12.01	34.04±11.17	36.60±16.62	36.19±10.11	32.09±14.53
CRP (mg/L)	4.62±2.78	4.51±2.68	5.10±3.49	5.05±2.77	4.00±2.81

NCS: Nutcracker Syndrome ANCS: Anterior Nutcracker Syndrome PNCS: Posterior Nutcracker Syndrome SD: Standart Deviation WBC: White Blood Cell MCV: Mean Corpuscular Volume MCH: Mean Corpuscular Hemoglobin MCHC: Mean Corpuscular Hemoglobin Concentration RDW: Red Cell Distribution Width MPV: Mean Platelet Volume NEU: Neutrophil LYM: Lymphocyte N/L: Neutrophil/Lymphocyte Ratio GLC: Glucose ALP: Alkaline Phosphatase ALT: Alanine Aminotransferase AST: Aspartate AminotransferaseLDH: Lactate dehydrogenase CK: Creatine kinase GGT: Gamma Glutamyl Transferase CRP: C-Reactive Protein

Table 2. Relation of symptoms snd signs with nutcracker syndrome type and gender

NCS	NCS Classification				Gender			
	ANCS		PNCS		Female		Male	
	Yes n (%)	No n (%)	Yes n (%)	No n (%)	Yes n (%)	No n (%)	Yes n (%)	No n (%)
Symptoms And Signs								
Side Pain	17 (63.0)	5 (18.5)	2 (7.4)	3 (11.1)	13 (48.1)	3 (11.1)	6 (22.2)	5 (18.5)
Abdominal Pain	15 (55.6)	7 (25.9)	4 (14.8)	1 (3.7)	14 (51.9)	2 (7.4)	5 (18.5)	6 (22.2)
Hematuria	11 (40.7)	11 (40.7)	0 (0)	5 (18.5)	8 (29.6)	8 (29.6)	3 (11.1)	8 (29.6)
Protoeniuria	10 (37.0)	12 (44.4)	0 (0)	5 (18.5)	7 (25.9)	9 (33.3)	3 (11.1)	8 (29.6)
HP	9 (33.3)	13 (48.1)	0 (0)	5 (18.5)	6 (22.2)	10 (37.0)	3 (11.1)	8 (29.6)
SPH	8 (29.6)	14 (51.9)	0 (0)	5 (18.5)	7 (25.9)	9 (33.3)	1 (3.7)	10 (37.0)
SPHP	7 (25.9)	15 (55.6)	0 (0)	5 (18.5)	6 (22.2)	10 (37.0)	1 (3.7)	10 (37.0)
Varicocele	3 (11.1)	19 (70.4)	1 (3.7)	4 (14.8)	0 (0)	16 (59.3)	3 (11.1)	8 (29.6)
Pelvic Congestion	4 (14.8)	18 (66.7)	1 (3.7)	4 (14.8)	5 (18.5)	11 (40.7)	0 (0)	11 (40.7)
Dyspareunia	7 (25.9)	15 (55.6)	1 (3.7)	4 (14.8)	6 (22.2)	10 (37.0)	2 (7.4)	9 (33.3)
Dysmenorrhea	10 (37.0)	12 (44.4)	1 (3.7)	4 (14.8)	11 (40.7)	5 (18.5)	0 (0)	11 (40.7)
Intolerance	4 (14.8)	18 (66.7)	0 (0)	5 (18.5)	4 (14.8)	12 (44.4)	0 (0)	11 (40.7)
Dyspepsia	11 (40.7)	11 (40.7)	1 (3.7)	4 (14.8)	8 (29.6)	8 (29.6)	4 (14.8)	7 (25.9)
Anorexia	6 (22.2)	16 (59.3)	0 (0)	5 (18.5)	5 (18.5)	11 (40.7)	1 (3.7)	10 (37.0)
Nausea	6 (22.2)	16 (59.3)	0 (0)	5 (18.5)	5 (18.5)	11 (40.7)	1 (3.7)	10 (37.0)
Vomitting	5 (18.5)	17 (63.0)	0 (0)	5 (18.5)	4 (14.8)	12 (44.4)	1 (3.7)	10 (37.0)
Weight Loss	6 (22.2)	16 (59.3)	0 (0)	5 (18.5)	5 (18.5)	11 (40.7)	1 (3.7)	10 (37.0)

NCS: Nutcracker Syndrome ANCS: Anterior Nutcracker Syndrome PNCS: Posterior Nutcracker Syndrome HP: Hematuria + Proteinuria SPH: Side Pain + Hematuria SPHP: Side Pain + Hematuria + Proteinuria

On imaging evaluation, 12 (44.4%) of the ANCS patients had RDUS, 13 (48.1%) had CT, 5 (18.5%) had MRI, 17 (63%) had RDUS and/or CT, and eight had both RDUS and CT, and none of the patients had retrograde venography (RV). While RDUS was performed in all five patients in the PNCS group, none of them had CT. As expected, imaging preference by gender was unrelated and rates were similar. Eleven (40.7%) patients in the ANCS group received conservative treatment, 6 (22.2%) received surgery, and 5 (18.5%) received endovascular therapy. Three (11.1%) patients received conservative treatment in the PNCS group, 1 (3.7%) received surgery, and 1 (3.7%) received endovascular treatment. Six (22.2%) females and 8 (29.6%) males were treated conservatively. Six (22.2%) female patients received endovascular therapy. Three (11.1%) male patients received surgical treatment, but no male patients received endovascular treatment (**Table 3**).

DISCUSSION

Only after alternative causes of hematuria have been ruled out is the diagnosis of NCS considered. The left renal vein is compressed between the aorta and superior mesenteric artery in the anterior NCS, whereas the retroaortic or circumferential renal vein is compressed between the aorta and vertebra in the posterior NCS. It is characterized by the clinical presentation of increased distal pressure. A rupture of thin-walled veins in the renal calyces is the most typical cause of hematuria (3). Other subtypes besides anterior and posterior have been defined. Nakazawa et al. (7) shown that the left-sided larger inferior vena cava compresses the left renal vein. Shah et al. (8) reported a case of left renal vein duplication in which the retro aortic branch was entrapped between the vertebral column and the aorta at the aortic bifurcation. Although the majority of cases

Table 3. Relationship of imaging-therapy status with nutcracker syndrome type and gender

NCS	NCS Classification				Gender			
	ANCS		PNCS		Female		Male	
	Yes n (%)	No n (%)	Yes n (%)	No n (%)	Yes n (%)	No n (%)	Yes n (%)	No n (%)
Imaging								
RDUS	12 (44.4)	10 (37.0)	5 (18.5)	0 (0)	11 (40.7)	5 (18.5)	6 (22.2)	5 (18.5)
CT	13 (48.1)	9 (33.3)	0 (0)	5 (18.5)	8 (29.6)	8 (29.6)	5 (18.5)	6 (22.2)
MRI	5 (18.5)	17 (63.0)	2 (7.4)	3 (11.1)	4 (14.8)	12 (44.4)	3 (11.1)	8 (29.6)
RV	0 (0)	22 (81.5)	3 (11.1)	2 (7.4)	1 (3.7)	15 (55.6)	2 (7.4)	9 (33.3)
RDUS CT	17 (63.0)	5 (18.5)	0 (0)	5 (18.5)	11 (40.7)	5 (18.5)	6 (22.2)	5 (18.5)
Treatment								
Conservative	11 (40.7)		3 (11.1)		6 (22.2)		8 (29.6)	
Surgical	6 (22.2)		1 (3.7)		4 (14.8)		3 (11.1)	
Endovascular	5 (18.5)		1 (3.7)		6 (22.2)		0 (0)	

NCS: Nutcracker Syndrome ANCS: Anterior Nutcracker Syndrome PNCS: Posterior Nutcracker Syndrome RDUS: Renal Doppler Ultrasonography CT: Computed Tomography MRI: Magnetic Resonans Imaging RV: Retrograd Venography

are anterior, it is also crucial to focus on the posterior group. 0.1% to 3.2% of left renal veins are circumaortic or retroaortic (9). Twenty-two of the twenty-seven participants in our study were anterior. Male patients exhibited higher posterior NCS than female individuals. This syndrome may exhibit symptoms in children, adolescents, and adults. In their second and third decades, men are diagnosed earlier than women (10). In a study involving pediatric patients, Çakıcı et al. (11) assessed 41 pediatric patients, of which 68.3% were female. In a separate study conducted by Orczyk et al. (10), 112 cases were analyzed, the distribution of males and females was determined to be equal, and the mean age at diagnosis for the entire group was 26.47±13.77 years. In our study, 16 (59%) of 27 patients were female, and their average age was determined to be 25.19±7.00 years.

The most common symptoms of the disease are hematuria and left flank pain. Hematuria can be observed macroscopically or microscopically. Macroscopic hematuria is associated with hypertension of the left renal vein. NCS should be investigated in patients with hematuria, proteinuria, left flank discomfort, and symptoms of pelvic congestion (12). In a study of 112 cases conducted by Orczyk et al. (10)., hematuria (78.57%), left flank pain (38.39%), proteinuria (30.36%), and anemia (13.39%) were the most common findings in patients. Besides, 35.71% of men had varicocele. Apart from anemia, hematuria, and proteinuria, no specific laboratory findings were observed in the cases in our study. Shin et al. (13) in a study conducted by the pediatric group, since the etiology was not found in 69% of the patients with hematuria, renal doppler ultrasonography was performed and NCS was detected in 40% of these patients. Microscopic hematuria is four times more common than macroscopic hematuria (13). The most common symptom was flank pain in 19 (70%) patients, followed by hematuria in 11 (41%) patients. While the prevalence of hematuria in our study was lower than that of comparable studies, the prevalence of flank pain was higher. Additionally, hematuria and proteinuria were common in patients with flank pain.

Similarly, orthostatic proteinuria occurs as a result of protein leakage from the calyceal system due to pressure increase. Hwang et al. (14) performed left renal venography and pressure evaluation of 23 children with orthostatic proteinuria, and showed that 12 (52%) cases had nutcracker syndrome. Although it is less common than hematuria, it can be seen in 0.6-10.7% (15). Proteinuria was observed in 10 (37%) patients in our case series, above the rates in similar studies. Shi et al. (16) identified 128 posterior NCS patients and showed that 22 of them had clinical findings, 11 had hematuria, and it was more prevalent in women than men, especially in the 18-40 age group. Even though posterior NCS was identified more frequently in male patients in our investigation, no hematuria nor proteinuria were detected in the posterior group. This indicates that the findings are less frequent in posterior NCS and that the diagnosis may be overlooked. In our study, patients in the anterior group had a higher prevalence of symptoms and findings, including dysmenorrhea, pelvic congestion, dyspareunia, dyspepsia, anorexia, nausea, vomiting, and weight loss.

Imaging is necessary in cases that require exclusion of other reasons for which the patient's clinic is compatible. Retrograde venography (phlebography) is the gold standard method for diagnostic accuracy because it evaluates both anatomy and pressure change. However, since it is invasive, it requires not routine practice in patients with poor clinical practice (17). Doppler US is preferred because it is a non-invasive diagnostic method. It provides information on both renal vein blood flow velocity and diameter measurement (18). In the studies conducted, important findings such as maximum diameter, flow velocity, and aortomesenteric angle were evaluated in terms of the diagnosis of NCS. Doppler US has disadvantages in terms of operator and patient cooperation. Another disadvantage in ultrasonography is to reveal a standard spectral doppler signal from the aortomesenteric segment of the left renal vein (19). In the studies conducted, it has been stated that the sensitivity of RDUS is between 69-90% and the specificity is between 89-100%. Kim et al (20) showed that it has 80% sensitivity and 94% specificity when the diameter and peak flow ratio measured by doppler US is greater than 5 in adult cases with NCS confirmed by venography. In our study, the most frequently used RDUS imaging technique was used on 17 patients. While no CT scan was available for five patients with posterior NCS, RDUS examination performed. We believe that with a good practitioner, RDUS can detect posterior NCS cases that are difficult to diagnose.

Since CT angiography does not cause invasive side effects (renal trauma, pseudoaneurysm) caused by retrograde venography, it is an important examination on the way to become the gold standard because of its rapid visualization of the renal vein and retroperitoneum with three-dimensional reconstruction (12). The narrowing of the diameter of the left renal vein in the aortomesenteric segment and the angle between the superior mesenteric artery and aorta below 41 degrees are the parameters that guide the diagnosis in axial tomography images. Also, MRI angiography can be preferred because it does not contain radiation, provides multi-planar

imaging, and shows the soft tissue anatomy in the compression area better. However, its transportation and use in practice are a little more difficult. However, CT angiography offers considerable diagnostic potential due to its greater accessibility. Shi et al. (16) scanned 6225 individuals, identified 128 patients with posterior NCS, and evaluated multi-slice spiral CT angiography as a viable non-invasive alternative to renal angiography for the study of renal vascular anatomy and variations. In our study, 13 individuals underwent CT and 7 experienced MRI. None of the patients in the posterior NCS group had a CT technique. But they all had RDUS. While there were no patients in the anterior NCS group who underwent retrograde venography, three of the five posterior NCS patients had RV. We think that this indicates the necessity of RV as an advanced invasive test for the diagnosis of posterior NCS. The absence of RV in any anterior NCS patients also suggests that advanced invasive examination may not be required in the diagnosis of anterior NCS.

Conservative, endovascular, laparoscopic, and surgical treatments are all possible treatment options. The treatment option may differ according on the clinic and renal vein pressure of the patient. Patients with moderate clinical conditions may utilize conservative therapy approaches. After 24 months of follow-up, patients with recurrent hematuria, significant flank discomfort, renal failure, and chronic orthostatic proteinuria must be referred to interventional treatment alternatives (21). Nephropexy, intravascular and extravascular stents, transposition of the left renal vein or superior mesenteric artery, gonadocaval bypass, renal autotransplantation, and nephrectomy are surgical procedures. In the pediatric age range, remission has been recorded to a considerable extent with growth and weight increase. Therefore, conservative treatment is the primary option (22). While surgery is the standard treatment, the recent success of endovascular interventions puts this option forward (23). Endovascular procedures have the disadvantages of necessitating long-term anticoagulant medication and the absence of information regarding long-term consequences (24). In 14 (61.8%) of the cases in our study, there was a correlation between conservative treatment and similar studies, suggesting that this approach is valid for the majority of patients. Eleven individuals with anterior NCS and two patients with posterior NCS received surgical and endovascular therapy. In addition, it is remarkable that only female patients had endovascular treatment in our study.

The most important limitation of the study was that it was retrospective, single-center. Another limitation was that the diagnosis studied was rare and could have been neglected.

CONCLUSION

NCS cases are frequently misdiagnosed or underdiagnosed because they are difficult to identify and need a complex diagnostic approach. In addition to patients who present with hematuria or flank pain in emergency or outpatient services, hematuric and proteinuric patients should also be investigated for the diagnosis of NCS. Considering this diagnosis, including in the differential diagnosis and planning the appropriate imaging method will contribute to the correct treatment process of the patients. Renal DUS can be used as an important non-invasive examination at both screening and diagnosis stages with a good practitioner. Tomography takes its place as a more risk-free diagnostic tool than venography. After early diagnosis, the success of conservative treatments is increasing. It can be said that non-invasive methods are gradually becoming more superior to invasive methods in both diagnosis and treatment processes.

ETHICAL DECLARATIONS

Ethics Committee Approval: The study was carried out with the permission of Bağcılar Training and Research Hospital Clinical Researches Ethics Committee (Date: 15.01.2021, Decision No: 2021.01.1.01.001).

Informed Consent: Because the study was designed retrospectively, no written informed consent form was obtained from patients.

Referee Evaluation Process: Externally peer-reviewed.

Conflict of Interest Statement: The authors have no conflicts of interest to declare.

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